

CHARACTERIZATION OF CHILDREN AND ADOLESCENTS WITH SICKLE CELL ANEMIA AND THE HEALTH SERVICES THEY USE¹

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ABSTRACT

The present study aimed to characterize children and adolescents with sickle cell anemia and to know the health services used by them. This is a descriptive study of a qualitative approach, carried out in a city located in the south of Brazil. The seven participants were legal guardians of children and adolescents with sickle cell anemia. The information was collected in the first semester of 2017 through a questionnaire filled out by the legal guardians. After the data collection, the information was analyzed through conventional content analysis. All the ethical precepts established by resolution no. 466/12 were respected. The results indicate that the most frequent complications of sickle cell anemia were vaso-occlusive crises, as well as infectious. In addition, participants have already gone through various health services to receive care, however, not all have accessed primary care. In view of the above, it is necessary to strengthen the network service articulating the various levels of complexity in health care and emphasizing primary care as a gateway.

Keywords: Sickle Cell. Anemia. Health Care. Pediatric Nursing. Child. Adolescent.

INTRODUCTION

The term sickle cell disease (SCD) is used for all hereditary disorders of hemoglobinopathies in which the clinical, hematological and pathological features are related to the presence of hemoglobin HbS in the blood. Meanwhile, the term sickle cell anemia (SCA) is used for one of the hemoglobinopathies in which normal hemoglobin (HbA) is partially or completely replaced by an abnormal hemoglobin (HbS), with a high complication rate, being the most severe form of SCD⁽¹⁾.

In view of this, it is important that SCD be detected early through neonatal screening, thus enabling the initiation of preventive measures leading to treatment, control of signs and symptoms, and acute and chronic complications, resulting in greater longevity and better quality of life. In this sense, the continuous monitoring of a health team is crucial⁽²⁾.

In recent years, the prognosis of this disease has improved, so individuals with SCA have received a life estimate until after the 50 years old. The highest risk of this disease occurs until the age of five, when the mortality rate is high, mainly due to infections.

However, sometimes, with proper treatment, children can live their daily lives normally, without the presence of symptoms; in adolescence, problems such as delayed growth and sexual maturation are common⁽¹⁾.

Chronic diseases are conditions that present periods of remission and exacerbation of symptoms, which may have a long or indefinite duration, with an often uncertain prognosis and requiring continuous care. Thus, these diseases require a new approach of care for services and health professionals, since these diseases are persistent or incurable and can accompany the individual throughout his/her life⁽³⁾.

In this context, the experiences of the children/adolescents must also be taken into account, because they can teach how to deal with the pathology. The disease becomes a constant companion for the patients, dictating behaviors and lifestyles, taking them to a restricted life, being a great part of their existence. Consequently, their lives is marked by social isolation, reduction of the dimension of work, of study and even of the family and by great material, physical and emotional investment⁽⁴⁾.

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In this way, in addition to guaranteeing access to health care for people with SCA, adequate and quality care must be provided. Quality in health care means the ability to offer human and personalized treatment that exceeds expectations regarding the fulfillment of their needs, whether they are explicit or implicit⁽⁵⁾.

Given the above and after identifying gaps in the publication of studies that present data referring to children and adolescents with sickle cell anemia, as well as the health services used by them, this study aimed to characterize children and adolescents with sickle cell anemia and to know the services they use. Thus, the guiding question is: What are the characteristics of children and adolescents with sickle cell anemia and what health services do they use?

METHODOLOGY

This is a descriptive study with a qualitative approach. Participants were located and invited in an outpatient service linked to a higher education institution in a city in the south of Brazil, where a group of patients with SCD are followed up through monthly meetings. The research participants were the legal guardians of five children and two adolescents with SCA, who participate in the said group.

Included in the study were those responsible for the children and adolescents with SCA followed in the aforementioned group. Those responsible for children and adolescents with sickle cell trait were excluded.

Data collection occurred from May to June 2017 through an interview with the legal guardians of children and adolescents with SCA at their homes. The questions were open and related to the identification of the child (age, sex, education and race - white, black, brown, yellow, indigenous or ignored) and to the clinical history (when the diagnosis was made; the medications used continuously; which complications the disease has already brought; whether blood transfusions have already been performed; whether any other treatment is used; what health services are used and how often). The interviews lasted an average of 20 minutes, were recorded on an audio device and transcribed manually.

After the data collection, the information was analyzed through conventional content analysis, in which the knowledge generated through this type of analysis is based on the individual perspectives of the participants and consists of real data⁽⁶⁾.

The research was carried out following the ethical assumptions contained in Resolution 466/12 of the National Health Council, which provide for ethics in research with human beings. To maintain the participants' anonymity, they were identified by characters from children's stories chosen by them. The information collection began after the approval of the research project by the Research Ethics Committee under the opinion number 1,973,446.

RESULTS AND DISCUSSION

The research participants were responsible for five children and two adolescents with SCA. Of the children, four were girls and one was a boy; the ages were in the age group of 8 to 12 years and they attended school, being in 2nd to 6th grades. The adolescents were 15 and 16 years old and attended the 8th and the 9th year of elementary school. As to the race of children and adolescents, three declared themselves black, two browns and two whites. As the disease originated in Africa and was brought to Brazil due to the large contingent of the African population that came to Brazil due to slave labor, its highest incidence is among blacks, but with the marked miscegenation in the Brazilian population, the disease has passed to affect other population groups⁽⁷⁾.

The clinical characteristics of children and adolescents, according to the questionnaire answered by the legal guardians, are presented in Table 1. It is highlighted, as shown in Table 1, that the children were diagnosed at birth by the neonatal screening (Bloodspot Test). On the other hand, the adolescents, due to the fact that the neonatal screening had not yet been implemented when they were born, were diagnosed through the hemoglobin electrophoresis test.

The Bloodspot Test consists of collecting blood drops from the child's heel to perform the neonatal screening tests. The collection should be performed between 48 hours after the first feeding of the newborn and the fifth day of life of the child, being one of the safest tests for the early detection. If the child did not perform this test in the first days after birth, there are other tests that are indicated for adults and children over four months of age⁽⁸⁾. Thus, the late diagnosis of the disease is intended for people of any age group. For this situation, the examination can be done by any of the existing methodologies for hemoglobin electrophoresis. The electrophoresis exam is in the Unified Health System (SUS) list and is part of the tests available in the Basic Health Units⁽⁸⁾.

Table 1. Clinical profile of research participants.

Identification	Diagnostic period	Diagnostic examination	Medications used	Complications occurred
Captain America	At birth	Bloodspot test	Folic Acid and Hydroxyurea	Pneumonia
Ariel	At birth	Bloodspot test	Carbamazepine and Folic Acid	Stroke and Splenic sequestration
Rapunzel	At three months	Bloodspot test	Folic Acid, penicillin, Tramadol, indication of hydroxyurea	Pneumonia, infections, painful crisis
Aurora	At six months	Bloodspot test	Folic Acid and Hydroxyurea	Splenic sequestration and painful crisis
Belle	At birth	Bloodspot test	Folic Acid and Hydroxyurea	Pneumonia and painful crisis
Wonder Woman	At birth	Bloodspot test	Folic Acid and Hydroxyurea	Risk of Stroke
Catwoman	At three years and six months	Hemoglobin Electrophoresis Test	Folic Acid and Hydroxyurea	Infection and painful crisis

After the diagnosis, the children should be referred, together with the parents, to a public health reference service for receiving specialized assistance, as well as guidance and information on the disease. Among the complications presented by the participants, the most common in this study is the vaso-occlusive crisis, which affects four participants, as well as infectious pictures. The vaso-occlusive crisis may appear due to sudden cooling of the skin and exposure to physical or emotional stress⁽³⁾. Infectious pictures occur in part because of the change in splenic function due to congestion and low blood flow. Vaccination and prophylactic use of antibiotics are important in reducing mortality, especially due to septicemia and meningitis⁽⁸⁾.

According to the collected information, all children and adolescents have already received blood transfusion, of whom two receive transfusions once a month, three from 10 to 20 transfusions, and two less than 10 transfusions. All children and adolescents had received a transfusion during hospitalization or were hospitalized to do so, three of whom have already received transfusions at the hematological center of the city and three had already sought emergency services to perform this procedure.

The complications of the pathology and the severity thereof are fears that are present in the daily routine of the family of the child and the adolescent with sickle cell anemia. This is the case of the family of Aurora, who lives with the complications of the disease and has to perform numerous blood transfusions. Every time the girl is hospitalized, the family suffers with the pains the child faces. There is also the fear that the pathology manifests itself in other children in the same family, as for the mother of Ariel,

who had recently given birth to a daughter and is afraid of the results of the tests and a diagnosis of SCD.

In the treatment of SCA, it is essential to resort to blood transfusions, but there are risks such as exposure to infectious agents that can lead to alloimmunization, blood hyperviscosity and hemosiderosis. Therefore, care should be taken and transfusions should be given only when indicated. In SCD, hemoglobin levels are as low as 6.0 g/dl, so anemia alone is not indicative of blood transfusion; one must rely on the basal hematological levels of each person and on the appearance of new signs and symptoms of hemodynamic decompensation⁽⁹⁾.

The health services used by children and adolescents are listed in Table 2. The table shows that all have access to specialized care, have episodes of hospitalizations and have already used the emergency service. Primary care is little sought, since only four of the seven children and adolescents had accessed this service. This shows a fragility in the service network, since it should have a link with users with chronic diseases in its coverage area.

The child and the adolescent with a chronic disease find little or no structure so that the care they need is comprehensive and takes place in an appropriate way to their demands, that is, beyond the hospital walls. This comprehensive care is still absent in an effective reference and counter-reference system for health care levels⁽¹⁰⁾. It is worth noting that the idea of networks is not something new in the Unified Health System. In the Federal Constitution of 1998, the article 198 already stated that health actions and services were part of a regionalized and hierarchical network, and primary care was considered the basis of the health system^(10,11).

Since SCD needs an early diagnosis and treatment to reduce the morbidity and mortality of children, the initial guidelines and the prophylactic and therapeutic treatment should be performed by a multidisciplinary team of primary care. However, this type of care is usually delegated to hematological centers⁽¹²⁾. Thus, intermediate levels of health care often ignore SCD, but they should improve care and reception, as well as insert these children and adolescents in the primary health care network, considering that this pathology is

a public health problem in the country⁽¹²⁾.

However, there is a recurrent idea of a network of care based on a strong centrality in its own logics of knowledge, making the user an object of action, as if they were devoid of knowledge and experience. When entering a health service it seems that the user needs to leave all the life that they bring from the outside, because in this meeting there is only room to reaffirm the already known, the way that the health professional considers 'correct' and which is the best way to live⁽¹¹⁾.

Table 2. Health services used by research participants

Identification	Hospitalization	Emergency services	Primary care	Specialized care
Captain America	X	X	X	X
Ariel	X	X		X
Rapunzel	X	X	X	X
Aurora	X	X		X
Belle	X	X		X
Wonder Woman	X	X	X	X
Catwoman	X	X	X	X

Thus, the public health system leaves gaps in the continuity of care, triggering consequences in the health actions aimed at this population. In addition, the fragility of the articulation of the health system and the ineffectiveness in the reference and counter-reference system, due to the lack of communication of the health network, are pointed out as limitations for the continuity of care^(13,14). These fragilities contribute to changes in the clinical picture of children and adolescents with chronic diseases and can lead to unnecessary hospitalizations, since the demands of this population are not met by this fragmented and specific care that has been offered by the public health system⁽³⁾.

Children and adolescents with a chronic disease need continuous care, reception and commitment from the professionals responsible for care. In addition, a network of care is also required for the various phases of the disease, and not only for the acute phase of the symptoms. This corroborates that the fragmented and disconnected organization of the health network, both at the programmatic level and at the local level, is unable to meet the care needs presented by this portion of the population⁽⁷⁾.

The care that children and adolescents with SCD need is not only in the hospital context, it does not die out at the time of hospital discharge, nor is it an exclusivity of high complexity services. There is a need for intra and intersectoral integration, in which it is possible to give continuity to care, so there is the

need of a resolute reference and counter-reference system, facilitating the flow of users within different levels of care, as well as qualified professionals in all these contexts⁽¹²⁾.

Many cities in Brazil do not have services able to provide health care to people with SCD, since many professionals do not have the necessary competence to provide comprehensive and adequate care for this population⁽¹⁵⁾. The professionals' lack of knowledge also causes them to minimize the necessary care when painful crises occur. Combined to this, the professional's difficulty in seeing the child and the adolescent as a comprehensive human being, targeting the technical part of care, considering only the biomedical model, hinders the adequate meeting of children and adolescents' needs⁽¹⁶⁾.

Due to the barriers that the health system imposes on users, such as prolonged waiting time and lack of contact with health professionals, the families of children and adolescents with sickle cell anemia feel unassisted by the primary care service, so they stop seeking this service. This is a great loss because, in addition to the follow-up of treatment, positive relationships between health professionals and users increase the desire to participate in consultations and reinforce the care that is necessary for children and adolescents with SCD⁽¹⁷⁾.

The health network and social support play a fundamental role in coping with the chronic condition of children and adolescents, and can mobilize

resources and people to meet the unique needs of this condition. In addition, health professionals need to participate in the formation of these networks in order to promote effective support for the maintenance of a qualified social environment for the child and the adolescent and their families, despite the adversities of the chronic disease, building and strengthening the potentialities and interfering positively in the experience of the supporters of these children and adolescents and in the care provided to them⁽¹⁸⁾.

For parents of children and adolescents of this study, the discovery of the pathology, their lack of knowledge about it, and the way health professionals passed on the information brought suffering and fear of the death of their children. This was the case of the mother of Belle, who upon receiving the diagnosis of her child, heard the prognosis that her daughter would survive until six months old, because the disease had no cure, according to information received from the health professional who gave her the results of the bloodspot test. A study of Ataíde and Ricas⁽¹⁹⁾ points out that the psychic suffering on the pathology is related to several changes and insecurities provoked by the diagnosis, among them the difficulty of understanding the disease due to the lack of knowledge and the lack of accurate information.

In this context, the health professional who makes the diagnosis should have the necessary knowledge about the disease and clearly communicate the information to the parents. The mother of Catwoman, whose diagnosis was made when the girl was three years old, told how difficult it was to hear from the doctor that her daughter would only live until she was five years old. Even 13 years after the discovery of the pathology, the mother is very frightened when Catwoman has some complication and the fear of losing her still ravages her. The mother of Captain America does not experience this feeling because she had access to more clarifying information when the boy was diagnosed with sickle cell anemia.

In addition, dealing with the pathology of the children also generates other difficulties for the parents, since the numerous hospitalizations impose leave from work and, consequently, financial difficulties. In this context, the use of medicines, such as Hydroxyurea, is also highlighted, which, if not provided by the government, becomes a monthly expense for the family. Rapunzel's mother had sent a request for this medication to the state government, and in the meantime, the girl ran out of medication because the family did not have the resources to buy it, which

generated a feeling of impotence in the parents regarding what they can do for the daughter.

For Wonder Woman's mother, the greatest difficulty was the daughter's coping with the disease; for this purpose, the mother sought support within the health services, but the lack of information about where to go and the difficulty to accessing the care network discouraged her to seek for a more quality care. Comprehensive care favors the minimization of hospitalizations and search for emergency services due to the monitoring and qualification of the health professionals involved in the care provided⁽²⁰⁾.

In this context, the training of health professionals and the mobilization of strategies that establish articulation and communication between the network would fundamentally qualify the care provided to children and adolescents with chronic diseases⁽³⁾. Therefore, the health team should focus on building and maintaining an appropriate relationship, as well as on creating strategies focused on the target public to improve clinical care⁽¹⁸⁾.

FINAL CONSIDERATIONS

Sickle cell anemia is a pathology that imposes several conditions on the lives of children and adolescents and their families, such as vaso-occlusive crises and infectious conditions, which interfere with adequate development, leading to withdrawal from the routine and the need for constant treatment. Many are the health demands of this population, and these lead to a pilgrimage between health services of different complexities. However, what was most evident in this study was the lack of integration in the health care network.

Primary care should represent, in all health conditions, the gateway to the health system. However, as in other situations, primary care is not accessed as a first option for the treatment of sickle cell anemia. In this sense, it is important to highlight the importance of articulating the services offered by the health system, in which primary care serves as a gateway and establishes connection with more complex services, such as specialized centers and hospitals.

Although the present study portrays a specific reality and has few participants for it has a qualitative character and does not aspire generalizations, we believe it may contribute to (re) thinking strategies for the care of children and adolescents with sickle cell anemia and their families. Therefore, it is imperative that health professionals who work at different levels

of complexity of the health system are able to provide comprehensive and resolute care.

CARACTERIZAÇÃO DE CRIANÇAS E ADOLESCENTES COM ANEMIA FALCIFORME E OS SERVIÇOS DE SAÚDE QUE UTILIZAM

RESUMO

Este estudo objetivou caracterizar as crianças e adolescentes com Anemia Falciforme e conhecer os serviços de saúde utilizados por eles. Trata-se de um estudo descritivo de abordagem qualitativa, realizado em um município situado no sul do Brasil, tendo como participantes sete responsáveis de crianças e adolescentes com Anemia Falciforme. A coleta das informações ocorreu no primeiro semestre de 2017, por meio de um questionário preenchido pelos responsáveis. Após a coleta as informações, foram analisadas através da análise de conteúdo convencional. Respeitaram-se todos os preceitos éticos estabelecidos pela resolução nº. 466/12. Os resultados indicam que as complicações mais frequentes da Anemia Falciforme foram as crises vaso-oclusivas, bem como os quadros infecciosos. Além disso, os participantes já passaram por diversos serviços de saúde para a continuação do cuidado a sua saúde, contudo nem todos acessaram a atenção básica. Diante do exposto, conclui-se que é necessário fortalecer o atendimento em rede articulando os diversos níveis de complexidade de atenção e enfatizando a atenção básica como porta de entrada.

Palavras-chave: Anemia Falciforme. Assistência à Saúde. Enfermagem Pediátrica. Criança. Adolescente.

CARACTERIZACIÓN DE NIÑOS Y ADOLESCENTES CON ANEMIA FALCIFORME Y LOS SERVICIOS DE SALUD QUE UTILIZAN

RESUMEN

Este estudio tuvo el objetivo de caracterizar a los niños y adolescentes con Anemia Falciforme y conocer los servicios de salud utilizados por ellos. Se trata de un estudio descriptivo de abordaje cualitativo, realizado en un municipio ubicado en el sur de Brasil, teniendo como participantes siete responsables de niños y adolescentes con Anemia Falciforme. La recolección de las informaciones ocurrió en el primer semestre de 2017, por medio de un cuestionario rellenado por los responsables. Tras la recolección, las informaciones fueron analizadas a través del análisis de contenido convencional. Se respetaron todos los preceptos éticos establecidos por la resolución nº. 466/12. Los resultados indican que las complicaciones más frecuentes de la Anemia Falciforme fueron las crisis vaso-oclusivas, así como los cuadros infecciosos. Además, los participantes ya pasaron por diversos servicios de salud para la continuación del cuidado a su salud, pero no todos accedieron la atención básica. Delante de lo expuesto, se concluye que es necesario fortalecer la atención en red, articulando los diversos niveles de complejidad de atención y enfatizando la atención básica como puerta de entrada.

Palabras clave: Anemia falciforme. Asistencia a la salud. Enfermería pediátrica. Niño. Adolescente.

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